Duplications of the Alimentary Tract

Clinical Characteristics, Preferred Treatment, and Associated Malformations

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Duplications of the alimentary tract are unusual congenital anomalies that frequently present a diagnostic as well as therapeutic challenge to the surgeon. Because these lesions occur so infrequently, they are often not suspected until encountered intraoperatively. Due to the complicated anatomy and common blood supply shared between the duplication and associated native bowel, appropriate management requires a familiarity with the anatomy and clinical characteristics of this entity. To better define the range of patient characteristics, clinical presentation, and preferred therapy, 20 enteric duplications were reviewed in 17 patients treated at the Children's Hospital Medical Center from 1956 to 1986. Ages of patients ranged from 1 day to 11 years; 60% were less than 2 years of age at initial presentation. Seven duplications in six patients involved alimentary tract structures of foregut derivation (esophagus, stomach, and Parts I and II of duodenum), with a predominance of girls (4 of 6). Most of these patients (67%) presented with moderate to severe acute respiratory distress and a mass present on chest radiograph. In 67% of the patients, the correct diagnosis was established before operation. None required emergency operative intervention. By contrast, 13 duplications in 11 patients were of midgut or hindgut derivation (Parts III and IV of the duodenum, jejunum, ileum, and colon). In this group of patients, 62% of the duplications involved the cecum, 23% involved the ileum, and 16%, the jejunum. Seventy-eight per cent of the patients were boys. The most common symptoms were nausea and vomiting, and the most common sign was a palpable abdominal mass. Emergency operative intervention was required of eight of 11 patients with duplications involving the small bowel and colon. Three patients presented with an intussusception, four with signs and symptoms consistent with acute appendicitis, one with a small bowel obstruction, and two with gastrointestinal hemorrhage due to the presence of ectopic gastric mucosa within the duplication. It was found that two important points must be considered in regard to the management of enteric duplications: (1) the common blood supply shared between the duplication and native bowel must be carefully protected to avoid undue sacrifice of normal bowel, and (2) the presence of heterotopic gastric mucosa in 35% of patients negates internal drainage. When resection of extensive tubular duplications would have required esophageal reconstruction or resulted in short bowel syndrome, selective mucosal excision was used in which the serosa and muscularis were left in continuity with the normal bowel and only the mucosa is removed, separating the duplication from the lumen of the associated alimentary tract structures.

DUPLICATIONS OF THE ALIMENTARY tract are rare. Most analyses have consisted of an isolated case report with an accompanying literature review. Before W. E. Ladd's pioneering analysis in 1937,[1] descriptive terms for this anomaly had included enteric or enterogenous cysts, giant diverticula, ileum, jejunum, or colon duplex, and "unusual" Meckel's diverticula. Ladd recommended that the term "alimentary tract duplications" be applied to those congenital malformations that involve the mesenteric side of the associated alimentary tract and share a common blood supply with the native bowel. His observations consolidated the classification of this entity and clarified its differentiation from other cystic malformations of the alimentary tract.[1]

The etiology of alimentary tract duplications has not yet been well characterized. Hypotheses have included the persistence of embryonic diverticula during development of the alimentary tract, intrauterine vascular accidents, and recanalization and fusion of embryologic longitudinal folds.[2,3] Abortive twinning has also been proposed as one possible causative factor of the extensive complete duplications of the colon and genitourinary system that occasionally occur.[4] However, all theories implicate the influence of an environmental stress that affects the development of the early fetus.

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This would explain the associated congenital abnormalities that are recognized as accompanying duplications of the alimentary tract.

The infrequency of alimentary tract duplications has limited analyses of patient characteristics, clinical presentation, and preferred therapy. These anomalies often mimic other disease entities and are most frequently encountered unexpectedly during laparotomy for what had been diagnosed as being a more common disease, such as intussusception, appendicitis, or gastrointestinal bleeding. To better define the clinical aspects of this anomaly, we have undertaken a retrospective analysis of 20 enteric duplications in 17 patients treated at our institution from 1956 to 1986. This relatively large series has allowed for the analysis of patient characteristics and preferred methods of treatment specific to duplications of the alimentary tract.

Patients and Methods

All patients with alimentary tract duplications treated exclusively at the Children's Hospital Medical Center from 1956 to 1986 were identified from medical records, surgical pathology reports, and operative reports. Patients with mesenteric cysts and those with duplications of the tracheobronchial tree were excluded. Twenty duplications in 17 patients were identified. There was a 100% complete follow-up for all patients.

Patient characteristics and symptomatology varied according to the embryological derivation of the involved tissues. Therefore, we analyzed the duplications in our series according to foregut, midgut, or hindgut derivation. Embryologically, the foregut at the cranial end of the primitive gut develops into the pharynx, respiratory tract, esophagus, stomach, first part and proximal half of the second part of the duodenum.[5] The midgut differentiates to form the distal half of the second part of the duodenum, Parts III and IV of the duodenum, the jejunum, ileum, cecum, appendix, and proximal colon to include the proximal two thirds of the transverse colon. The hindgut differentiates to form the distal one third of the transverse colon, the descending and sigmoid colon, the rectum, anus, and a significant part of the urogenital system.[5] Because duplications of midgut and hindgut derivation were so similar in clinical characteristics, we have combined them for further analysis.

Results

Patient Characteristics

Twenty alimentary tract duplications were identified in 17 patients treated at our institution from 1956 to 1986. Patients ranged in age from 1 day to 11 years at initial presentation; 60% were less than 2 years of age at the time of diagnosis of the first duplication. Overall, there was a slight predominance of boys (60%).

Thirty-nine per cent of duplications involved alimentary tract structures of foregut derivation (esophagus, stomach, and Parts I and proximal half of Part II of the duodenum), and 61% were of midgut or hindgut origin (distal duodenum, jejunum, ileum, cecum, and colon). Overall, the most frequent site was the ileocecal area (8 of 20).

Alimentary Tract Duplications of Foregut Derivation

In six patients, seven duplications involved alimentary tract structures of foregut derivation. Six of the foregut duplications involved the esophagus, and one, which was a second noncontiguous duplication, involved the greater curvature of the stomach. There was a predominance of girls among these patients (4 of 6). Four of these six patients were less than 2 months of age at initial presentation. The majority of the patients (67%) presented with moderate to severe acute respiratory distress, and all had a mass present on chest radiograph. In one patient, a mass was noted on routine chest radiograph obtained for evaluation of aseptic meningitis. In another patient, a mass was noted in the upper right thoracic area. In all patients, a barium swallow with upper gastrointestinal series confirmed the mass. A representative study is shown in Figure I. In 67% of the patients, the correct diagnosis was established before surgery.

Characteristics Specific to Duplications of the Esophagus

There were six esophageal duplications in our series. Because esophageal duplications have attendant considerations specific to this anatomic site, we have analyzed this foregut location separately. Two of the six were extensive and tubular in nature. Both of these were in communication with the native esophagus, and mucosal excision with preservation of the seromuscular layers was successfully performed. The cystic esophageal duplications were easily resected by complete excision. A representative cystic duplication is illustrated in Figures 2A and B.

The histology present in esophageal duplications was variable (Table 1). All duplications contained at least two types of epithelium, including transitional, columnar, heterotopic lung tissue, thyroid stroma, ganglia, and various squamous varieties. Two of the six esophageal duplications contained parietal glands with heterotopic gastric mucosa, and one contained well differentiated lymphoid aggregates resembling Peyer's patches.

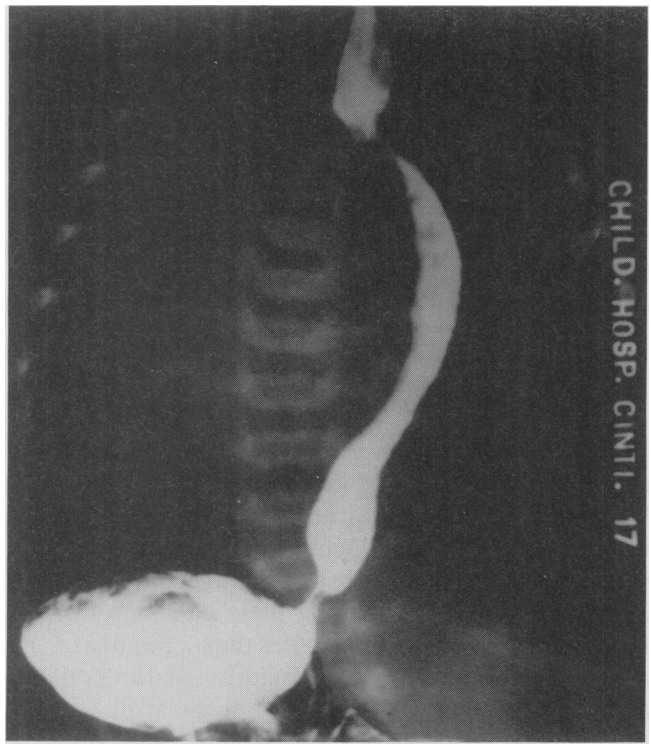


FIG. 1. Radiograph of an esophageal duplication, demonstrating displacement of the esophagus by the duplication.

Three of the six patients with esophageal duplications had associated congenital malformations: two with vertebral anomalies including cervical or thoracic hemivertebrae, and one with idiopathic scoliosis and metatarsus adductus. Two of these three patients had a second noncontiguous alimentary tract duplication. Both patients with foregut duplications and a second alimentary tract duplication had an associated congenital malformation.

TABLE 1. *Histology of Ectopic Tissue Present in Duplications of the Esophagus*

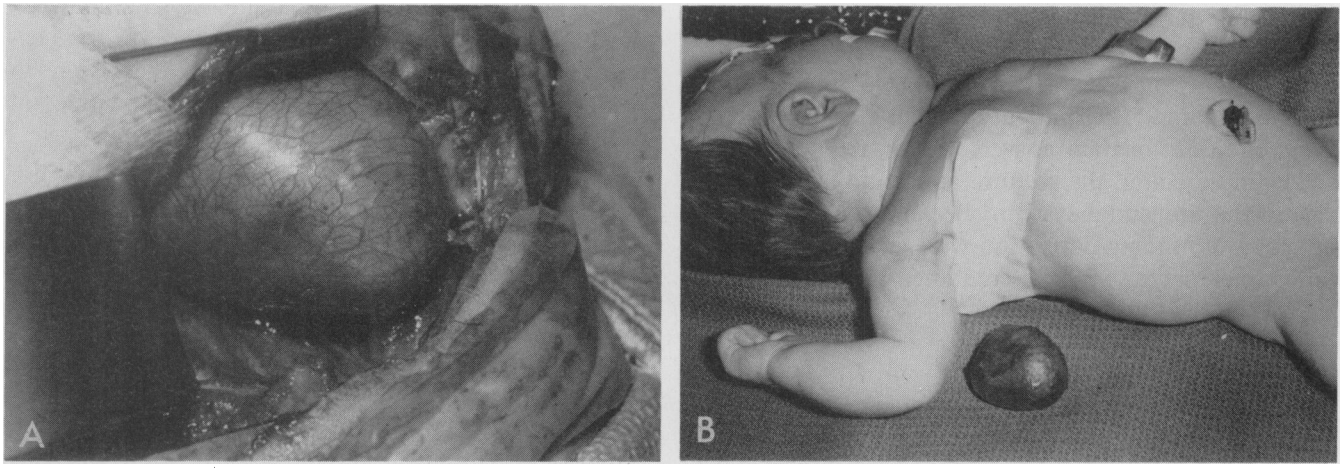
Patient Number	Epithelium	Gastric Mucosa	Peyer's Patches
E1	Columnar Transitional	No	No
E2	Mucous glands Small bowel villi	Yes	Yes
E3	Heterotopic lung Nasopharyngeal glands Submucosal glands	No	No
E4	Mucous columnar cells Ganglia, squamous	Yes	No
E5	Squamous, ciliated Cuboidal cells Mucous glands	No	No
E6	Thyroid stroma Respiratory muscle Squamous epithelium	No	No

Both second duplications were below the diaphragm; one involved the greater curvature of the stomach, and the second involved the proximal ileum.

Alimentary Tract Duplications of Midgut and Hindgut Derivation

Thirteen duplications in 11 patients were of midgut or hindgut embryologic derivation (Parts III and IV of duodenum, jejunum, ileum, cecum, colon, rectum, and anus). Seventy-eight per cent of these patients were boys. In this group of patients, 62% of duplications involved the ileocecal area, 23%, the jejunum, and 16%, the ileum. Representative surgical specimens are shown in Figures 3, 4A, and 4B. One duplication was a complete duplication of the colon, including two appendixes.

In contrast with patients who had duplications of foregut origin, the preoperative diagnosis was less readily established in patients with duplications of mid-



FIGS. 2A and B. (A) Operative photograph of duplication exposed through an extrapleural right thoracotomy. (B) Photograph of duplication immediately following its removal. The common muscular wall between the duplication cyst and the esophagus was left on the esophageal side, removing only the mucosa of the duplication.

gut and hindgut derivation. Three patients presented with an intussusception with the duplication as the intussusceptum, four with signs and symptoms consistent with acute appendicitis, one with a small bowel obstruction, and two with significant gastrointestinal hemorrhage secondary to the presence of ectopic gastric mucosa. In one patient with a second esophageal duplication on upper gastrointestinal x-ray series, a duplication was identified before surgery. In only three of the 13 patients was the correct diagnosis established before operation. Emergency operative intervention was required for eight of the 11 patients.

Treatment consisted of resection with primary reanastomosis in all patients except two, who had extensive tubular duplications involving the small bowel (Fig. 5). In these two cases, mucosal excision was used with preservation of the seromuscular layer of the duplication in continuity with the native bowel, resulting in preservation of the blood supply to the native bowel.

All duplications of midgut and hindgut origin had at

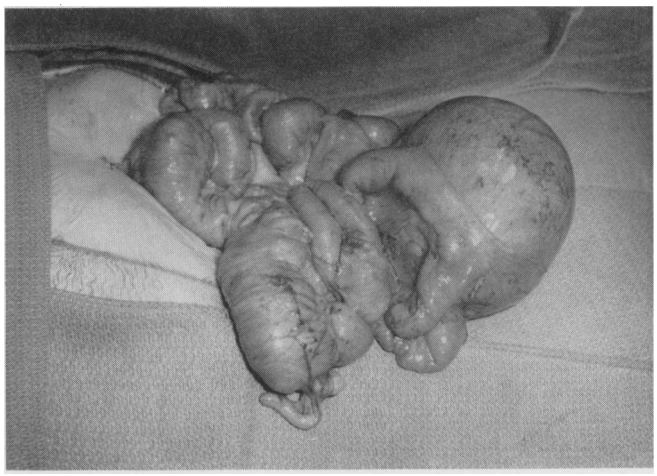
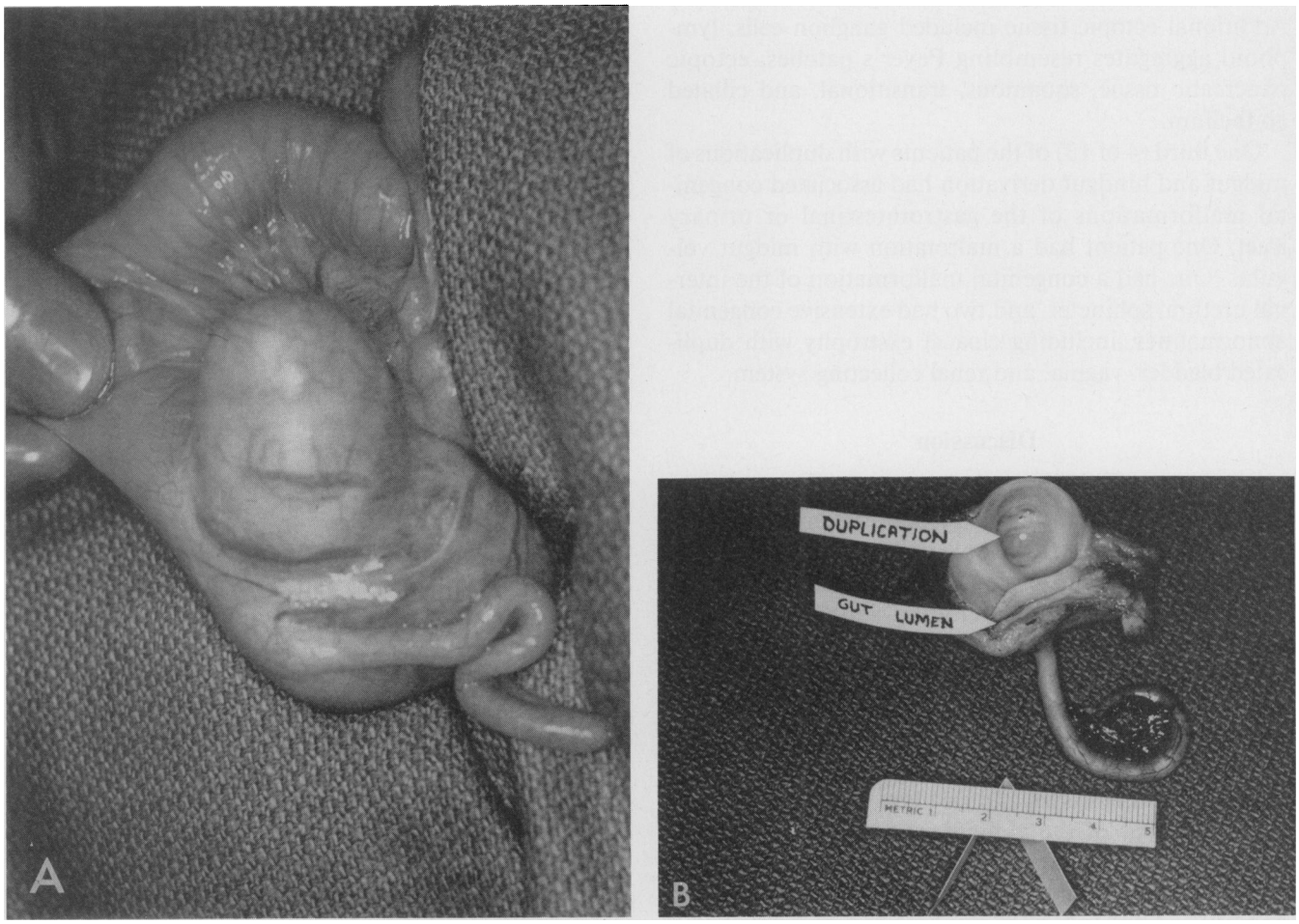


FIG. 3. Operative photograph of duodenal duplication showing second portion of duodenum coursing over the surface of the mass. The duplication was intimately associated with the pancreas, pancreatic ducts, and blood supply to the duodenum. Amylase content of the duplication was elevated, indicating that it received drainage from the pancreas.



FIGS. 4A and B. (A) A cystic duplication of the small intestine that did not communicate with the lumen of the bowel. (B) Specimen opened to demonstrate the lumen of the duplication and lumen of the adjacent terminal ileum.

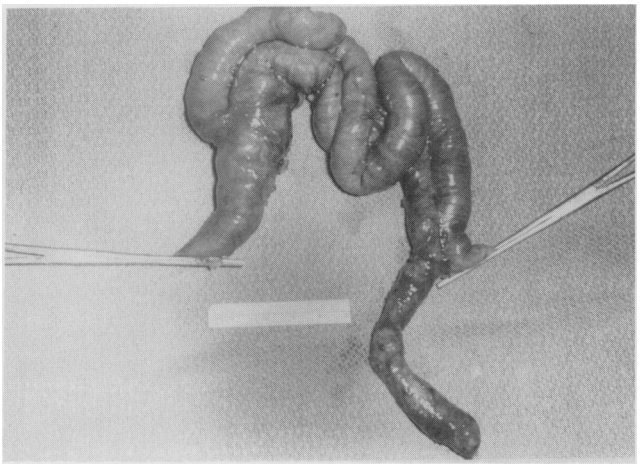


FIG. 5. Photograph of a tubular duplication of the small intestine in which the blood supply went first to the duplication, then to the small intestine. This was treated by resection and end-to-end anastomosis. For a more extensive tubular duplication, an alternate approach would be selective mucosal excision to remove the mucosa only, leaving the seromuscular layers of the wall and the blood supply intact.

least two types of epithelial tissue present. In five of the 13 duplications, ectopic gastric mucosa was present. Additional ectopic tissue included ganglion cells, lymphoid aggregates resembling Peyer's patches, ectopic pancreatic tissue, squamous, transitional, and ciliated epithelium.

One third (4 of 13) of the patients with duplications of midgut and hindgut derivation had associated congenital malformations of the gastrointestinal or urinary tract. One patient had a malrotation with midgut volvulus. One had a congenital malformation of the internal urethral sphincter, and two had extensive congenital abnormalities, including cloacal exstrophy with duplicated bladder, vagina, and renal collecting system.

Discussion

Multiple hypotheses have been proposed to explain the etiology of duplications of the alimentary tract.[2-4] The presence of heterotopic tissue, including well formed circular and/or longitudinal muscular layers, and lining epithelium, including squamous, transitional, and tubular and acinar glands, is supportive of a congenital developmental origin. In our series, there was commonly a presence of heterotopic tissue of diverse varieties: thyroid stroma, gastric mucosa, lymphoid aggregates resembling Peyer's patches, ciliated bronchial epithelium, lung tissue, and cartilage.

Duplications of the alimentary tract were most frequent in the ileocecal region in our series. Because of the intra-abdominal nature of these abnormalities, in the majority of patients with duplications of midgut and hindgut derivation, the diagnosis was established intraoperatively and was not suspected before operation. Patients presented with intussusception, signs and symptoms consistent with acute appendicitis, bowel obstruction, and gastrointestinal hemorrhage due to the presence of ectopic gastric mucosa. By contrast, the majority of duplications of foregut embryology were suspected before surgery, due to the uniform presence of a mass on chest radiograph and the results obtained from contrast radiographic studies that suggested the diagnosis. Most patients with esophageal duplications had symptoms referable to the respiratory tract, and all had a mass that was present on chest radiograph.

Duplications of the alimentary tract may be tubular or cystic in configuration,[4] with the cystic form being the more frequent in our series. Both types were characteristically located on the mesenteric aspect of the associated native bowel, and therefore shared a common blood supply.[4] Those of tubular configuration tended to be more extensive in our experience.

Operative management of enteric duplications requires a familiarity with the entity and may pose a challenge to even the most skilled surgeon. Two important points must be considered: (1) the common blood supply shared between the duplication and the native bowel must be carefully protected to avoid undue sacrifice of normal bowel, and (2) the presence of heterotopic gastric mucosa negates internal drainage because of the risk of gastrointestinal hemorrhage from this focus. In our series, 35% (7 of 20) of duplications contained heterotopic gastric mucosa, and two of those patients initially presented with gastrointestinal hemorrhage.

Generally, cystic duplications were easily removed from their attachments to the surrounding tissues. Some were literally "shelled-out". Others required operative resection with primary end-to-end anastomosis to restore bowel continuity. Those of tubular configuration were more extensive, and at times posed a special challenge to the surgeon. When extensive resection would have resulted in short bowel syndrome or have required esophageal reconstruction, we found selective mucosal excision to be an excellent alternative approach. With this technique, the serosa and muscularis are left in continuity with the native bowel, and only the mucosa of the duplication is removed, separating the duplication from the lumen of the associated normal alimentary tract structures. In this manner, extensive resections were avoided and all heterotopic tissue excised. This technique was first reported by Wrenn in 1962,[6] and we have subsequently used it in four patients. In patients treated in this manner, the gastric mucosa did not regenerate.

Congenital anomalies which accompany alimentary tract duplications may be explained in part by the embryological origin of the structures: foregut, midgut, or

hindgut. Half of our patients with duplications of foregut derivation had associated skeletal malformations including thoracic and cervical hemivertebrae, idiopathic scoliosis, and metatarsus adductus. By contrast, one third of the patients with duplications of midgut and hindgut derivation had congenital malformations involving the gastrointestinal or genitourinary tracts without associated skeletal abnormalities. All of the patients in our series with a second noncontiguous alimentary tract duplication had an associated congenital malformation of the skeletal, gastrointestinal, or genitourinary tract. We therefore recommend for all patients with a documented duplication and an additional congenital malformation that the surgeon look for an additional noncontiguous duplication.

In conclusion, although duplications of the alimentary tract are rare, appropriate management requires a familiarity with the anatomy and clinical characteristics of this entity. Patients with duplications in our series presented with a variety of nonspecific symptoms and signs, often requiring urgent operative intervention. Alimentary tract duplications must be considered during the evaluation of thoracic and abdominal masses, gastrointestinal hemorrhage of unclear etiology, intussusception, and mechanical bowel obstructions. The common blood supply shared by the duplication and associated normal bowel must be recognized and preserved in order to avoid unnecessary and catastrophic loss of normal bowel. Our experience indicates that those patients with an alimentary tract duplication plus an associated malformation of the skeletal, gastrointestinal, or genitourinary tract should be thoroughly evaluated for a second noncontiguous alimentary tract duplication. Finally, internal drainage is not an option for therapy because of the relatively high (35%) incidence of ectopic gastric mucosa present within the lining mucosa of the duplications.

Acknowledgments

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